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Lady windermere syndrome variant: Atypical presentation in young female patient

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Abstract
Non tuberculous lung infections with Mycobacterium are rare in this part of the continent. Lady Windermere syndrome (LWS) refers to the appearance of bronchiectasis, centrilobular nodules leading to the lung scarring in elderly population. This results in volume loss and the right middle lobe and lingual lobes are the mostly affected. The lung changes are the result of pulmonary Mycobaterium avium complex (MAC).We present a 24-years old young female who presented with the history of cough with off and on haemoptysis with breathlessness. Plain chest radiograph had shown some scar in the right mid zone. High resolution computerized tomography (HRCT) confirmed the findings of underlying bronchiectasis and collapse of right middle lobe. This was further confirmed by culture of bronchalveolar lavage fluid for MAC. The patient was put on combination with clathyromycin; rifampin and ethambitol for two years with regular follow up. The patient had shown remarkable improvement for the last one year.

Keywords: non tuberculous; Lady Windermere syndrome; bronchiectasis; MAC; bronchoalveolar lavage

Introduction
MAC infections takes place in the elderly females who are in habit of suppression their cough and expectoration. This group is other than Mycobacterium tuberculosis and Mycobacterium leprae [1, 2].This leads to the changes in right middle lobe or lingular lobe on left side. The term was coined in 1992 by Reich and Johnson. This syndrome is named after a fastidious character in one of Oscar Wilde's plays "Lady Windermere's Fan" [3].

Case report
24-years old immunocompetent female from the rural background reported to the pulmonology outpatient department with complaints of cough, expectoration and with off and on haemoptysis of two years duration. She was nonsmoker and without any history of any systemic illness. She had the habit of suppression of the cough in the public. The environment of the residential place was also polluted because of the vehicular traffic in the nearby road.Patient was tall and thin female. On examination she was found to be afebrile,with sinus rhythm without tachypnoea. Skin tests with allergens were unremarkable. IgG, IgM, IgA, IgE and alpha-1 antitrypsan levels were normal. Systemic examination was unremarkable. Plain chest radiograph had shown some linear opacity in the right middle zone (Figure 1).

Fig 1: Plain chest anteroposterior radiograph.Ther is partial collapse of right middle lobe which is seen in the form of linear opacity (white horizontal arrow) with the secondary signs of collapse,

The patient was subjected to high resolution computerized tomography (HRCT) of the chest. This had shown the collapse of right middle lobe with underlying bronchiectatic changes (Figure 2a and b and Figure 3). There were multiple pretracheal lymph nodes with central necrosis (Figure 4a and b).

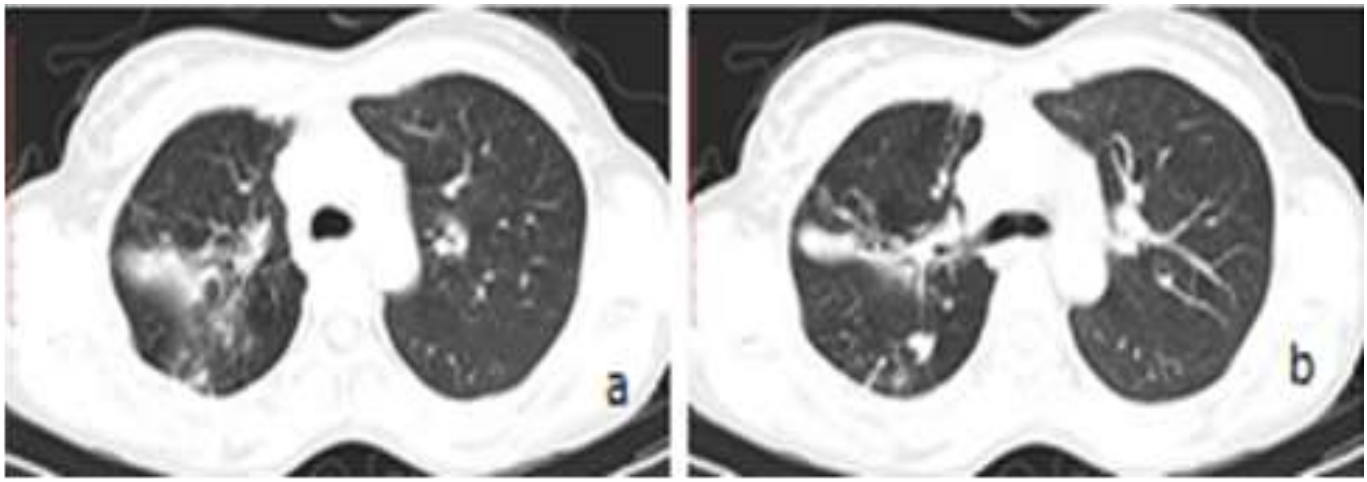

Fig 2: HRCT Chest.a) upper part of the right middle lobe with evidence of inhomogenous opacity with decrease in lung volume.b) axial section just below the bifurcation of trachea showing plate like atelectasis.

Fig 3: HRCT contd.Magnified axial view of the part of the lung showing underlying bronchiectatic changes in the right middle lobe.

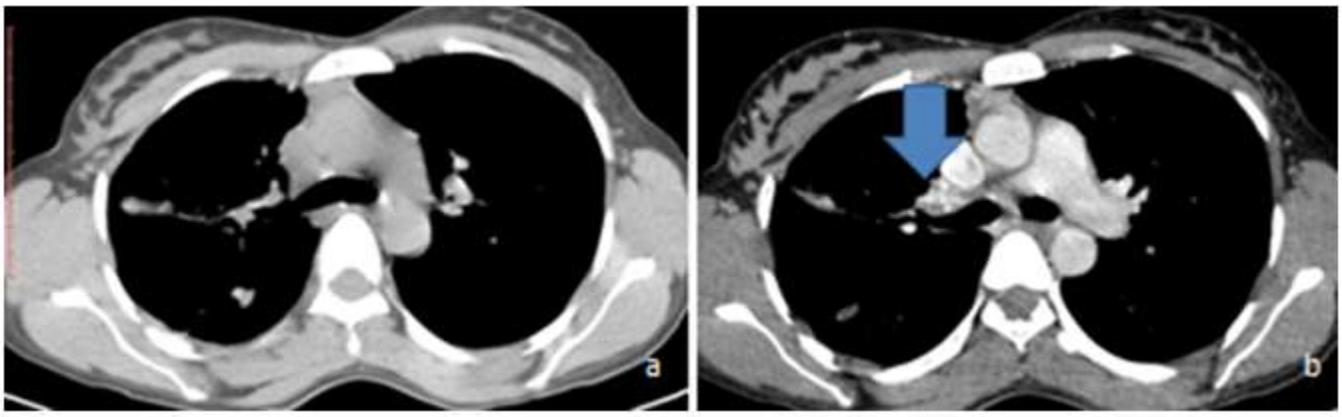

Fig 4: CT Chest mediastinal window. a) NCCT chest axial section shows the atelectasis patch. b) CECT chest shows multiple lymph nodes on right paratracheal region (inverted arrow)

Bronchoscopy was done and there was no evidence of any endobronchial lesion or obliteration of the right middle bronchiole.Brochoalveolar lavage fluid smear did not show any mycobacterium bacillus. The culture had shown the confirmation of Mycobacterium avium cellularis.The patient was put on the combination of clathyromycin, rifampin and ethambitol for two years with regular follow up. The patient had shown remarkable improvement without any ill effects for the last one year and still on monthly follow up.

Discussion

Non tuberculous infections did not draw much attention before the entry of HIV infections.These differ in many ways of symptomatology and manifeastations [4,5].There has to be some preexisting lung disease.Thes can be divided into following three: categories:

a. When preexisting pulmonary ailment
b. Non-preexisting lung ailment
c. atypical presentation

Our present case present in atypical category as the patient was neither an immunocompromised neither with the previous lung disease. The presence of these type of cases are very rare in Indian population as per literature [7].LWS is more common in those females who had no history of smoking or pulmonary disease. They are usually thin built. The bronchiectatic pattern is of nodular type. The distribution of the bronchopulmonary tree of right middle and left lingular lobes are considered to play mechanism of this entity (Figure).The bronchii are small in caliber without collateral ventilation The hypothesis is because of their long distribution and acute angulations the stasis of secretion is more in this part of the lung.. [8].Those who voluntarily suppresses the cough are the individuals of MAC category.

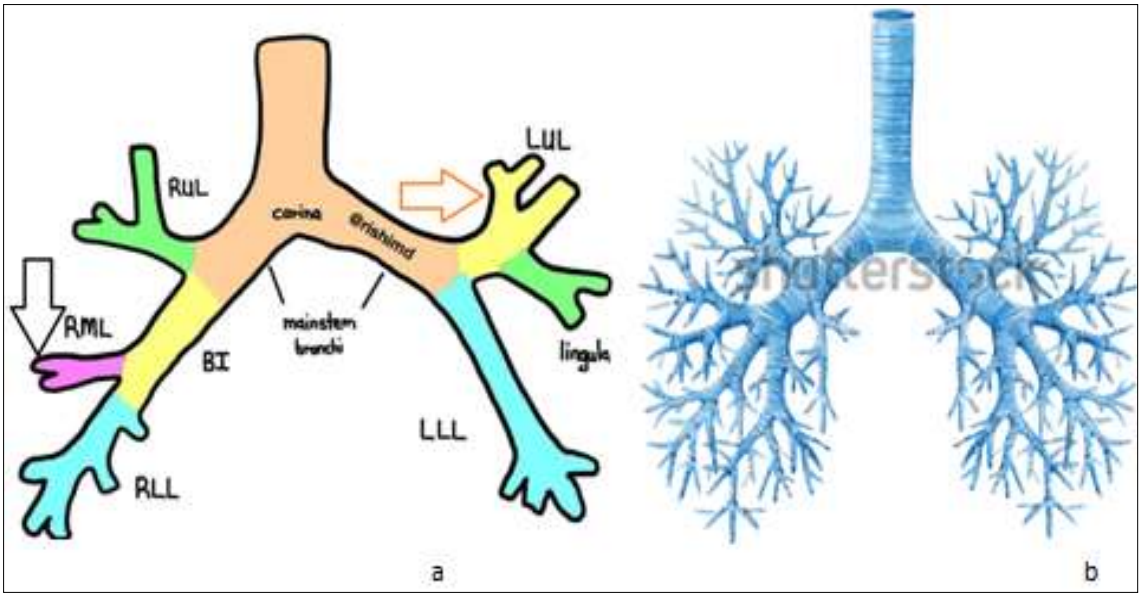

Fig 5: Diagrammatic description of bronchial tree. a) in anteroposterior view with distribution as per lobes. b) the same profile is seen as a complete tracheobronhial tree. There is long and angulated tree distribution of right middle lobe (inverted white arrow) and left lingular lobes (horizontal red arrow).

Other individuals who fall in this group are to suppress the cough because of the sterna pain or discomfort which they suffer

because of frequent coughing [9]. Iseman *et al* correlated LWS patients with the underlying skeletal abnormalities like scoliosis, pectus excavatum or straight back [10]. Some cases may be related to the underlying connective tissue disorders but our case does not fall in any of the category [8].

Conclusion

LWS cases are usually seen in elderly females who are in habit of voluntarily suppression of the cough. This leads to accumulation of secretions of the already bronchiectatic lung. This leads to the collapse of either right middle lobe or lingular segment of the left lobe. This is usually caused by MAC.

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Consent of the patient

The written consent of the patient was taken

Conflict of interest

The authors have no conflicts of interests or funding sources to disclose

References

1. Field SK, Fisher D, Cowie RL. Mycobacterium avium complex disease in patients without HIV infection. Chest. 2004; 124:466-581.
2. Kim RD, Greenberg DE, Ehrmantraut ME, Guide SV, Ding L, Shea Y, *et al*. Pulmonary nontuberculous mycobacterial disease:prospective study of a preexisting syndrome.Am J Respir Crit Care Med. 2006; 178:1066-1074
3. Glassroth J. Pulmonary disease due to nontuberculous mycobacteria. Chest. 2008; 133(1):243-251
4. Reich JM, Johnson RE. Mycobacterium avium complex pulmonary infection presenting as isolated lingular or middle lobe pattern:the lady Windermere Syndrome. Chest. 1992; 101:1605-9.
5. Manglani RP, Khaja M, Hennessey K, Kennedy O. Pleural mycobacterium avium complex infection in an immunocompetent female with no risk factors.Case Resp Pulmonol. 2015; 2015:760614.
6. Isseman MD, Buschman DI, Ackerson IM. Pectus excavatum and scoliosis: thoracic anomalies associated with pulmonary disease caused by Mycobacterium avium comlex. Am Rev Respir Dis. 1991; 144:914-16.
7. Iseman MD. Mycobacterium avium complex and the normal host.N Eng J Med. 321:896-8.
8. Byrd RP, Mirza S, Roy TM. Lady Windermere's syndrome:an atypical presentation of an atypical mycobacterium,J Tenn Med Assoc. 1995; 88:303-5.
9. Hutchings HA, Morris S, Eccles R, Jawad MSM. Voluntary suppression of cpough induced by inhalation of capsaicin in healthy volunteers.Respir Med. 1993; 87:379-82.
10. Prince DS, Peterson DD, Steiner RM. Infection with Mycobacterium avium complex in patients without predisposing conditions. N Eng J Med. 1989; 321:863-8.